IV

CONGENITAL SYPHILIS

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THE attempt is sometimes made to classify syphilis as it occurs in children into two distinct categories—*congenital* and *hereditary*. According to some, congenital syphilis is said to occur when symptoms of the disease are present at birth; while in hereditary syphilis these do not manifest themselves until some time has elapsed. Others would confine the latter term to instances of infection of the ovum from the maternal blood, or from the paternal spermatozöon. It seems to the writer that not only is such a distinction unnecessary, but that it is impossible; and, furthermore, that it is scientifically wrong to use the term "hereditary" with reference to syphilis.

This matter is purely a biological one, and it is very important in the interests of clear thinking that biological terms should be used in their correct sense when dealing with disease and the problems associated with it.

By "heredity" is meant the organic or genetic relationship between successive generations—between offspring and parent. The term "inheritance" expresses all that the organism is, or has, to start with, by virtue of its hereditary relation. These characters have their initial seat in the fertilised ovum—the physical basis of inheritance being found in the germ-cells.

From a biological point of view diseases may be classified under two heads:—

(1) Abnormal processes springing from germinal peculiarities which express themselves during development.

(2) Abnormal processes directly induced by some extra-germinal or environmental factor, such as a micro-organism.

Syphilis comes under the second head.

A micro-organism can never be part of the inheritance, even were it possible for it to exist in the germ-cells. It

cannot form part of the organisation of the germ-plasm. Ante-natal infection is not inheritance. A child may be born with syphilodermata ; it may be born in apparent health and develop interstitial keratitis some years afterwards ; but all that these things indicate is that intra-uterine *contagion* has occurred. The disease has been *acquired* from the parent during the period of gestation ; and this is an entirely different thing from inheritance. There is no essential difference between a child born with syphilis and an adult acquiring a chancre. The former has been infected before birth, and the latter after. It is a grave misunderstanding of biological terms to apply the labels " hereditary " or " inherited " to any specific infective disease with which a child may be born. It can only cause confusion.

Syphilis, then, may be congenital—a child may be born with it whether or not superficial signs be present ; it can never be hereditary. The common error is to regard biological inheritance as being of the same nature as a material legacy, such as an estate. There is really no such parallelism. At first the organism and the inheritance are identical. They are one and the same thing. The inheritance can only express itself under the influence of nurture, that is, during the process of development. This is in no way comparable to an individual and the property and title to which he succeeds. Such an one merely acquires the ancestral estate and the earldom by virtue of the genetic relationship between himself and some other individual.

It has been thought expedient to consider this matter of nomenclature since a vast amount of unprofitable—sometimes acrimonious—discussion in respect of scientific matters has arisen because the terms used have not been properly defined, or have been employed in a loose fashion.

Congenital syphilis, then, may be defined as syphilis which is acquired *in utero*, and which may or may not manifest itself at birth. The rare cases in which a child is infected during its passage through the birth-canal are simply instances of *acquired infantile syphilis*. The only practical point here is, that in congenital syphilis the infection is a massive one ; in syphilis which is acquired after birth this is not so. In the former case the child enters the world with an enormous syphilitic burden. Such a child is a " saturated solution of syphilis." In

non-congenital cases the patient can never reach the point of syphilitic saturation.

The diagnosis of congenital syphilis commences with the parents. While it is important that a syphilitic person should not marry, it is equally important that where syphilis exists in either or both marital partners, conception should not be allowed to take place. Should such an undesirable event occur, the pregnancy will surely terminate in abortion, miscarriage, still-birth, or in a congenitally syphilitic child. When the incidence and ubiquity of syphilis is thoroughly appreciated a serological test—provocative if necessary—will, in every case of pregnancy, become as much a routine procedure as is the examination of the urine for albumen at the present day. Not until this happens is the national scandal of 30,000 annual still-births and deaths from syphilis of children under the age of one year likely to cease. The incidence of congenital syphilis will reach the vanishing point only when every pregnant woman who has, or whose husband has, syphilis, is treated from the commencement of her pregnancy up till her confinement.

The Public Health aspect of this matter is of vital national importance. It is no exaggeration to say that with a Maternity Service and a Venereal Diseases Scheme working under the same local authority, the number of syphilitic children that exist is a blot upon the escutcheon of British preventive medicine. One is of the opinion that every case of congenital syphilis can be diagnosed before birth, and that with intensive treatment from the early days of pregnancy a healthy child can invariably be produced. The Wasserman surveys which have been carried out in various parts of the country among pregnant women, especially those of the hospital class in large cities, clearly indicate that, from the point of view of preventive medicine, the possibility of syphilis should be considered in every instance. All that is required is an awakening of the Public Health conscience of the general practitioner and a closer *liaison* between the Maternity and Venereal Disease Departments controlled by the Medical Officer of Health. In maternity work the routine of instilling protargol or something similar into the eyes of the new-born has almost abolished ophthalmia neonatorum ; the routine Wassermann would go a very long way towards stamping out congenital syphilis.

Some time ago there was published by the Medical Research Council a monograph on "Maternal Syphilis as a Cause of Death of the Fœtus and of the New-born Child." With most of the conclusions therein arrived at one finds oneself in profound disagreement, and, since these are apt to influence opinion as to prevention, diagnosis and treatment of syphilis in the mother and child, one hopes upon a future occasion to review this monograph and to indicate where one thinks the author is in error. In the meantime one would venture to enunciate the following principles :—

(1) If a child is born whose blood, or whose mother's blood, gives a positive Wassermann test, that child should be regarded as suffering from congenital syphilis. The positive Wassermann constitutes an indication for treatment in spite of the absence of clinical signs.

(2) A child whose blood at birth is negative, born of parents of whom one or both give positive tests in the blood or cerebro-spinal fluid, or in whom these have been positive at any time during the two years preceding the pregnancy, and who have had inadequate or no treatment, should be regarded as a congenital syphilitic. By suitable provocation the blood of such a child will give a positive result.

(3) If the parents of a child born with a negative blood can be proved to have been free from syphilis for the two years preceding the pregnancy, or if the mother has been under intensive treatment throughout the period of gestation, the child may be considered free from disease and treatment withheld. It is prudent, however, to perform provocative serological tests at intervals during the first two years of life. If such a child develops any obscure symptoms, in spite of the negative tests, anti-syphilitic treatment should be instituted.

(4) The treatment of the child of syphilitic parents begins on the day pregnancy is diagnosed.

After birth the diagnosis of congenital syphilis is not always so simple a matter as is sometimes supposed. Sins of omission are far too frequent. Many of these are undoubtedly due to too much reliance being placed upon

the negative result of an unprovoked blood-Wassermann test. Experience shows that frequently, in spite of a history of abortions and miscarriages, in spite of the discovery of former syphilis in the parents, sometimes in spite of the presence of a positive blood in the mother, treatment is withheld because the child appears healthy and its blood is negative.

It is important to bear in mind that at birth all the organs and tissues of the congenital syphilitic are affected, and that a stage is apparently reached very rapidly which is comparable to the endosyphilitic stage in acquired syphilis. The disease becomes concentrated in certain organs and tissues, giving rise either to no symptoms whatever, or only to those of such a vague character that they are not diagnostic of syphilis. The unprovoked Wassermann in such instances is, as a rule, negative.

The more remote effects of congenital syphilis are of very high importance, and it is essential that they should be recognised. It would seem that comparatively few cases of congenital syphilis exhibit the classical stigmata of the disease. Leredde [1] shows that mental retardation in children is frequently due to syphilis, and that in such cases excellent results follow upon antisyphilitic treatment. He recommends that in all early cases of mental retardation such a procedure should be adopted. The same writer [2] describes three cases of epilepsy of recent origin in which there were no other signs of syphilis. These were all cured by anti-syphilitic treatment. One case was in a child of ten years who had *petit mal* for three months. The parents were syphilitic. Since treatment the child has remained well. The other two were cases of *grand mal*, and after treatment for syphilis there have been no further attacks. During the last three years one has seen four cases of typical general paresis in children, all of which had been labelled " epilepsy." At the present day it ought not to be asking too much that in all cases of infantile nervous disease an investigation should be made to rule out the possibility of syphilis.

Kemp and Poole [3] have made an interesting investigation into familial neurosyphilis. Twenty families were taken, each of which possessed one or more children suffering from congenital neurosyphilis. These were compared with another group of twenty families, the children of which were congenital syphilitics, but in whom the

disease did not affect the central nervous system. It was found that neurosyphilis was eight times more frequent among the mothers and three times more frequent among the fathers of the former group than of the latter. These findings would seem to indicate either that there is a neurotropic strain of treponema pallidum, or that there is in the first group an inherited predisposition to disease of the central nervous system. In the United States, the division of the Public Health Service dealing with venereal diseases has published in "Social Pathology" some interesting statistics with regard to the prevalence of congenital syphilis among juvenile delinquents. The general conclusion arrived at is that delinquency is twice as common among congenital syphilitics as among healthy children. It seems that the congenital syphilitic is a psychopathic individual with an extremely unstable nervous equilibrium. If such an one is not specially treated and guarded against becoming a delinquent, he will inevitably tend towards anti-social acts.

Chatenet [4] in a series of seventy-five children suffering from late congenital syphilis, whose ages ranged from two to fifteen years, found that in 84 per cent. there were present certain signs not usually regarded as diagnostic of the disease. He comes to the conclusion that a history of miscarriages, premature births, a family history of high infantile mortality, hydramnios, melæna neonatorum, convulsions, retarded mental and physical development, strabismus, early rickets, craniotabes, enlarged thymus, jaundice, unilateral hydrocele, are extremely suggestive of congenital syphilis. In his series there were rarely seen any of the classical signs, and any of the syphilides were uncommon. He is of the opinion that the manifestation of any of the above symptoms in children is an indication for the immediate institution of anti-syphilitic treatment irrespective of what may be the result of a Wassermann test.

One is being forced to the conclusion that a negative unprovoked Wassermann test on the blood of infants in no way eliminates the possibility of syphilis. Some time ago, Åhman [5] drew attention to the frequency with which clinical signs of syphilis may exist in infants while the Wassermann test is negative. It is common to find that a negative result obtained during the first few days of life may soon become positive, even when anti-syphilitic

treatment has been instituted. It is important also to realise that in children, the blood may remain negative for a very long time during which there may be abundant clinical evidence of the activity of the disease. There would appear, however, to be little doubt that at certain times and by suitable provocation syphilis can be demonstrated serologically in the majority of, if not in all, cases. There is much that is still obscure in the serology of congenital syphilis ; in the present state of our knowledge we must base our procedures upon parental history and upon the presence of the conditions mentioned by Chatenet. It may not be a highly scientific method, but practically the therapeutic test is of immense value ; and especially in cases of infantile nervous diseases.

" Epilepsy " may be caused by syphilitic osteitis of the cranial bones. Leri and Cottenot [6] report three such cases in which the fits were of the Jacksonian type. In one patient there was a gumma of the skull, in another there were syphilitic erosions, and in the third hyperostosis. In each instance anti-syphilitic treatment resulted in rapid improvement. A similar case is described by Louste and Louet [7] as occurring in a man aged twenty-three who had recurrent attacks of Jacksonian epilepsy for two years. Radiologically there was discovered cranial hyperostosis in the right temporo-parietal region. The Wassermann test was positive, Hutchinsonian teeth were present, and signs of aortitis were found in the mother.

Syphilis manifests itself in more diverse ways in the congenital than in any other form of the disease ; and it is too often forgotten that the classical stigmata are in no way evidential of the present activity of the treponema pallidum. The peg-shaped and notched teeth, the saddle nose, and the fissures at the angles of the mouth are permanent. They persist after the patient is cured. The signs of active congenital syphilis are often obscure, and at times but little information is to be gained by ordinary serological investigation. In the obscure troubles of infancy, instead of syphilis being thought of last, it should be considered first. In spite of a negative Wassermann the therapeutic test should be applied, the pregnancy-record of the mother should be inquired into, and the serological reactions of the parents should be explored.

Blum and Fatou [8] publish a report of six cases of late

congenital syphilis of the joints. Some of these were not of the osteo-articular type, but were purely synovial in character. Such cases are apt to simulate acute rheumatism, being accompanied by great pain and a rise of temperature. The synovial fluid is sterile and shows from 70 to 95 per cent. of polymorphonuclear leucocytes. Bilateral hydrarthrosis is strongly suggestive of congenital syphilis. In all the cases under review a positive Wassermann test was recorded.

Bretervide and Bretervide [9] draw attention to the importance of the radiological examination of the aorta as a means of diagnosing congenital syphilis. Increased calibre and a shadow denser than normal they consider to be absolute evidence of syphilis, whatever may be the serological findings. This conclusion is based upon a series of 462 children. The normal maximal diameter of the aorta is 1 cm. up to eight years of age, and 1·3 cm. from eight to fourteen. They also find that an exaggerated second sound at the left margin of the sternum in the second and third interspaces is present. It is probable that many of the aortic lesions found in adult life have had their origin in early childhood, when, had they been discovered and treated, cure would have resulted.

The treatment of congenital syphilis is by no means always easy; and it is, of necessity, prolonged. The first fact to realise is that the little patient is saturated with the disease. His organs and tissues are, as a rule, swarming with treponemata. By too intensive treatment at the beginning there is a very real danger that the rapid killing off of large numbers of the organisms may cause a dangerous degree of toxæmia. It is for this reason that one has of late been more sparing in the use of arsenicals, and has begun the treatment with bismuth. A few cases of new-born children have been treated by bismuth inunctions, and although the series is at present too small upon which to dogmatise, one has gained the impression that the results obtained are somewhat better than those from mercurial rubbings. One is of the opinion that congenital syphilis cannot be cured in under five years, and that it is prudent to continue the treatment up to puberty. The scheme that one adopts at present in a new-born child for its first year of life is :—

Bismuth inunction for one month.
Arsenic intramuscularly for two months.

Bismuth intramuscularly for three months.
Arsenic intramuscularly for three months.
Bismuth intramuscularly for two months.
Iodides orally for one month.

For each of the succeeding four years the patient receives two courses of treatment, each extending over a period of four months. Once the diagnosis of congenital syphilis has been made, the duration of treatment is not gauged by the serological reaction. It is not a case of chasing a negative Wassermann. Five years is fixed as the minimum, and treatment is continued for that time, as outlined above. If then the Wassermann is negative, treatment is discontinued; if positive, it is persevered with.

REFERENCES

(1) LEREDDE. *Bull. Soc. franç. de Dermat. et Syph.*, 1924, xxxi., 294.
(2) LEREDDE. *Bull. Soc. franç. de Dermat. et Syph.*, 1924, xxxi., 237.
(3) KEMP and POOLE. *Jour. Amer. Med. Assoc.*, 1925, lxxxiv., 1395.
(4) CHATENET. *Thèse de Paris*, 1925. No. 492.
(5) AHMAN. *Hygiea*, 1924, October 31st, 725.
(6) LERI and COTTENOT. *Bull. Soc. franç. de Dermat. et Syph.*, 1924, xxxi., 242.
(7) LOUSTE and LOUET. *Bull. Soc. franç. de Dermat. et Syph.*, 1924, xxxi., 409.
(8) BLUM and FATOU. *Presse Méd.*, October 14th, 1925, 1366.
(9) BRETERVIDE and BRETERVIDE. *Arch. de Méd. des Enf.*, 1924, xxvii., 257.